Ano Rectal Malformations – A Study

A three year study - Aug2008 to July 2011

A Dissertation submitted in partial fulfillment of M.Ch Branch V (Paediatric Surgery) examination of Dr. M. G. R. Medical University, Tamil Nadu, Chennai, to be held in August 2011.

Certificate

Certified that the dissertation – entitled "Ano Rectal Malformations – A Study" is the Bonafide work under taken by Dr. M. SHANKAR under my guidance and supervision, in the Department of Paediatric Surgery, Government Rajaji Hospital, Madurai Medical College, Madurai, during the period of his Postgraduate residency in M. Ch. Paediatric Surgery from 2008 to 2011.

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DECLARATION

I, Dr. **M. SHANKAR** solemnly declare that this dissertation "**Ano Rectal Malformations – A Study**" was prepared by me under the guidance and supervision of Professor and HOD, Department of Paediatric Surgery, Madurai Medical College and Government Rajaji Hospital, Madurai between 2008 and 2011.

This is submitted to The Tamil Nadu Dr. M.G.R. Medical University, Chennai, in partial fulfillment of the requirement for the award of **MASTER OF CHIRURGIE, in PAEDIATRIC SURGERY,** degree Examination to be held in **AUGUST 2011.**

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Date : **Dr. M. SHANKAR**

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Dr. M. Shankar, GRH, Madurai, 2011.

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Introduction

Ano Rectal Malformations (ARM) represents a complex group of congenital anomalies resulting from abnormal development of the hindgut, Allantois and Mullerian duct, leading to incomplete or partial Uro rectal septal malformations.

ARM is a relatively common congenital cause of intestinal dysmorphology in the newborn. There are epidemiological differences in the level and extent of the abnormality. The spectrum of lesions varies from fairly minor lesions (e.g. Covered Anus) to some of the most complicated and complex abnormalities Cloaca, Exstrophy and Rectal atresia. The defect may include many systems - e.g. Curarino's Triad includes CNS and Vertebral Defects, Exstrophy includes bladder defects. These are a few of the myriad of presentations. It is one of the most complicated defects to correct and forms the back bone of Paediatric Surgery, the details of which is the aspiration of every student applying himself to this field.

ARM forms a significant load on the surgical services, particularly in developing countries, not only in the emergency situation but also in terms of long-term corrective procedures. Although there have been major advances in the management of these children during the last 15 years, these patients still represent a continuing challenge as a result of the significant reconstructive problems involved, as well as the fact that a significant number suffer from faecal and urinary incontinence, as well as the possibility of inadequacies -

sexual, non correctable defects, not to talk of the associated anomalies in later life.

With development in the Surgical Specialities the management has improved and what was a certain disaster has now been converted to normal livelihood, and we now see patients who have married and borne children with a normal life span.

In our study we have mainly stressed on the demographic profile of the disease and the bearing and inferences which we can aggregate from them, which have been well interpolated towards the end along with the appraisal of the surgical procedures.

Review of Literature

Low socio-economic status

Congenital defects have been associated with various harmful agents to which the mothers were exposed during the critical period of embryogenesis, thalidomide being a good example. In a previous study penile agenesis and congenital sacrococcygeal teratoma in the population were linked with the frequent use of insecticides. The affected population in the present survey was largely of low socio-economic status and were most likely to burn and inhale mosquito repellant coils at night or to fumigate their farms with insecticides. These agents may affect pregnant mothers, resulting in the possible birth of malformed babies. Also, it has been documented that teenage pregnancy is a major problem in our society and in attempts by these young mothers to terminate their pregnancies they ingest various concoctions. Presumably, the constituents of some of these medicaments have teratogenic effects. This hypothesis constitutes a subject for further research. Poverty and ignorance were noted to be the main factors affecting treatment outcome.[1] A concerted public enlightenment campaign is therefore required.

High Protein diet, Folic and Iron Supplementation has a salubrious effort in preventing ARM in our scenario. This has to be stressed on a war footing.

Psychosocial burden and poor quality of life

The quality of life of the parents was also affected. The expressed feelings were areas for concern. Predominant complaints were feelings of despair, anxiety, depression, futility of life, difficulty in access to health services, in adequate improper information provided to them, the means of transport, and their financial conditions rather than the ethical social aspects and treatment of the child. The last two concerns are important considering the low socioeconomic status of most parents in the present study. This resulted in discontinuity of treatment or improper attendance to hospitals. More intensive family support and motivation may be indicated for those with these individuals – Family members of child afflicted with ARM. Though a minority group they have these common impediments - Low socioeconomic status, limited social support, or high perceived burden. The present study concludes that there is greater psychosocial burden and poor quality of life among parents of children with ARM. It is imperative to provide psychosocial support, including promotion of a clear understanding of the disease to these participants.[2]

Insurance companies if really desiring improvement of quality of life of patients should look into this matter and see how the families afflicted by congenital anomalies could be refurbished.

Associated malformations

In a study by G. R. Boocock and D. Donnai, One hundred and sixty nine patients with anorectal malformation were studied. There were 108 boys, 60 girls, and one case of intersex. Low malformations were more common in both sexes. Over half the subjects had associated malformations. These were more common in the group with high malformations. There was no difference between the sexes in this respect. A family history of similar malformation was found in 15 cases (9%). Where anorectal malformation was the only abnormality in the family an autosomal dominant mode of inheritance was likely, except in one case where there was consanguinity. Where there were associated malformations no single mode of inheritance emerged. Multiple associated malformations may indicate recessive inheritance and subsequent pregnancies should be regarded as high risk and full antenatal investigative facilities provided.[3]

In a study by Boemer et al., it is recommended that all patients with anorectal malformations should have all necessary investigations to search the associated anomalies different systems. However large number of patients and poor primary health care services make us confine to do basic investigations rather than follow a protocol. So we routinely do Ultrasonography of abdomen, X-ray spine of all patients. Special investigations for example: Intravenous Urography, MCUG and fistulogram are done in appropriate cases. Actual incidence of Uro Genital anomalies may be higher if thoroughly investigated,

with specialized investigations like CT scan, MRI Urogram, etc., on a routine basis.[4]

Smith ED, Saek. M. N. 1988 stated that Plain X ray chest, abdomen and pelvis can help to diagnose associated anomalies like Vertebral anomalies, Sacral anomalies, Cardiac anomalies, Esophageal atresia with or without fistula, Duodenal atresia and other bowel atresias like small bowel or colonic atresia. Cystic termination of the gut, complications like spontaneous bowel perforation due to volvulus of sigmoid (distension and gangrene), Iatrogenic perforation due to perineal exploration in a high anomaly may be noted by these basic studies.[5]

Embryology

Kluth D, Lambercht W states that exact mechanism of development of anorectal is still mysterious and controversial. Two main events are thought to be important for differentiation of cloaca. The conventional view is that down growth of the Urogenital septum divides cloaca into Urogenital (anterior) and anorectal (posterior) parts. Controversy does still persist whether the division takes place by downward frontal septum (Tourneux fold), median fusion of lateral folds of Rathke or a combination of both. The cloacal membrane is divided into anal and Uro genital membranes.[6,7]

The hindgut has a ventral diverticulum called allantois. The dilated cavity receiving the hindgut proper and allantois is lined by endoderm and is called endodermal cloaca. Ventrally endodermal cloaca is closed by cloacal

membrane. Endodermal cloaca with hindgut proper and allantois are together surrounded by mesenchyme. At the junction of the hindgut and allantois there is proliferation of the mesenchyme and endoderm, and as a result a septum develops called Uro Rectal septum.

Classically the Uro Rectal septum is formed by down growth of Tourneux's fold and ingrowth of the lateral folds of Rathke that fuses in the midline. The septum divides the endodermal cloaca into the dorsal part, which develops into rectum and anal canal and the ventral part, which develops into vesico urethral part and urogenital sinus. The septum also divides the cloacal membrane into the posterior part called the anal membrane and the anterior part called the urogenital membrane. Perineal body develops at the junction of the two membranes.[6]

Chatterjee and Roy proposed that internal cloaca is separated from the external cloaca by cloacal membrane. The majority of mal formations are due to the growth failure of the hindgut or agenesis. Arrest of growth takes place at various levels giving rise to high, intermediate or low malformations.

Classification

In 1970 at symposium on Anorectal malformation at the pediatric surgical congress in Melbourne based on work done by Smith and Stephens,[8] the international classification system was proposed .

Type of Anomaly	Female	Male
High	1. Anorectal agenesis A: Rectal atresia B: With fistula Rectocloacal fistula Rectovaginal /high 2. Rectal atresia	1. Anorectal agenesis A: Rectal atresia Rectovesical fistula Rectourethral fistula 2. Rectal atresia
Intermediate	1. Anal agenesis A. Without fistula B. With fistula Rectovaginal fistula low Rectovestibular fistula 2. Anorectal stenosis	1. Anal agenesis A. Without fistula B. With fistula Rectobulbar fistula 2. Anorectal stenosis
Low	1. At normal anal site Covered anus – complete Covered anal stenosis 2. At perineal site Anocutaneous fistula Anterior perineal anus 3. At vulvar site Vulvar anus Anovulvar fistula Anovestibular fistula	1. At normal anal site Covered anus – complete Covered anal stenosis 2. At perineal site Anocutaneous fistula Anterior perineal anus
Miscellaneous	Anal membrane stenosis Imperforated anal membrane Perineal groove Perineal canal	Anal membrane stenosis Imperforated anal membrane Perineal groove Perineal canal

Other classifications are

- Ladd and Gross classification 1934.[9]
- Stephens and Smith1963 classification based on embryological concepts.
- Anomalies based on a simplified Santulli classification.[10]
- Wingspread Conference classification. [11]
- Krickenbeck Classification.

Radiology in Anorectal Malformations

Wangensteen and rice[12] in 1930 found that Invertogram will not be useful when the bowel shadow does not reach the distal pouch in situations like esophageal atresia, duodenal atresia or small bowel atresias. Over distended small bowel shadows also can give a deceptive picture in the invertogram. Accurate interpretation of invertogram of the pelvis requires some thought and planning. The important precautions to be taken while doing an invertogram are: Invertogram should be taken after 12 to 18 hours of life to allow enough time for adequate bowel gas to pass to the end of the blind rectum. If the bowel is not overly distended with air one may wait 4-6 hours before placing a nasogastric tube for gastric decompression that will permit enough air to pass down to the gastrointestinal tract. But one should guard against over distension of bowels because unrelieved massive bowel distension producing sigmoid volvulus and spontaneous bowel perforation can occur in some babies by 24 hours of life.

Baby should be held vertically upside down for 1 minute before the film is taken. The baby should be quite and should not cry or move during exposure. The hips should be slightly extended or kept relatively straight so that the femur do not obscure the pubic bone. It is very important to obtain a true lateral view of the pelvis where both the right and left ischium overlie each other exactly and accurate centering on the greater trochanter. Placing a thin smear of barium

paste in the buttocks cleft at the level of the external sphincter helps to denote the cutaneous level of the anus.[12]

In 1973, a prone crosstable radiograph was recommended as an alternate to the classic invertogram for diagnosing the level of rectal atresia. The baby is placed in Prone Jack-knife position for a few minutes. A lateral film is taken centering on the greater trochanter. An advantage of prone lateral x rays are easy to perform and gives superior radiographs. Baby is not disturbed much during the procedure so baby can rest quietly in this position for a longer period. The chances of aspiration during the procedure are less especially in babies with Tracheo esophageal fistulae. Dr. K. L. N. Rao, is credited for this lateral shoot in lieu of Invertogram.

Interpretation of invertogram and prone lateral x rays

This is based on the relationship of the air in the distal blind pouch to the pubococcygeal line and the 'i' point. Pubococcygeal line is the line drawn from the upper border of the symphysis pubis to the sacrococcygeal junction. 'P' point is centre of the boomerang shape of the Os pubis and C' point is just caudal to the last (fifth) ossification centre of sacrum. Pubococcygeal line passes through the upper cresentic margin of the ossified ischium. The cranial one quarter with caudal three quarters of the ischial shadow. Pubococcygeal line in babies with sacral agenesis can be developed by projection from the pubis through the same site on the ischium which serves as a reliable alternative

landmark. In these circumstances the 'PC' line lies well caudal to the last ossified vertebra of the defective sacrum. Ischial line ('l' line) is drawn through the 'I' point parallel to the pubococcygeal line; 'I' point is the inferior end of ischial comma.

In male bladder neck, verumontanum and anterior peritoneal reflection of the rectum are at 'pc' line. In female external os of the cervix is located at 'pc' line. Bulb of urethra in male is located at the level of ischial line. The urethral orifice is lies caudal to the "I' point. In Anal agenesis and in rectourethral fistula and gas shadow reaches the ischial line and this is the lowest point of the levator in these deformities. In female the 'I' line corresponds to the upper limit of the perineal body and the level of triangular ligament.

Interpretation of invertogram :

In high or supra levator anomaly the blind pouch ends at or above the pubococcygeal line. In intermediate anomalies are in the rectum ends between pubococcygeal line and ischial point. In low or translevator anomaly air in the blind pouch is below the 'I' line.

There are several fallacies in the interpretation of invertogram. Gas shadow at a much higher level than expected may be due to contraction of the puborectalis muscle while taking pictures. Gas shadow may not be smooth and rounded due to active contraction of the puborectalis or meconium in the distal rectum. Holding the baby in the inverted position may succeed in displacing the

meconium. Gas shadow at a level lower than expected may be due to the child straining excessively or due to excessive pressure on the abdomen.

Some intermediate lesions will appear to have a gas shadow below the 'I' point, occasionally in a male child with an intermediate anomaly with Rectobulbar fistula the pouch may be filled with gas and lie below the 'i' point thus simulating a translevator anomaly.

In female babies with vaginal fistula the invertogram may not reveal the level of blind pouch because of escape of gas and meconium through the fistula. Presence of a 'Beak' anteriorly may identify the level of the fistula. But the 'Beak' may be present in some cases even without fistula. Gas in the vagina or a low lying loop of small bowel may mimick a low lesion. Invertogram may show air in the bladder which in a female indicate rectovesical fistula and in a male may be rectovesical or rectourethral fistula.[12]

Murugasu et al and Carmin et al in 1972 described percutaneous injection of soluble contrast material through the perineum into the distal pouch may be useful in demonstrating the distal end of the bowel and in outlining a fistulous connection to the genitor-urinary tract. Direct injection of contrast through a fistula or anus is useful in defining the level of an anorectal stenosis, anovestibular or rectovestibular fistula and useful in distinguishing between rectobulbar and rectourethral fistula. This information can be obtained from voiding cystourethrography or retrograde urethrography.[13]

Shopfner CE in 1965 demonstrated that micturating cystourethrography is performed at an early stage which may find the site if any of a rectourethral or anourethral fistula. Even if the fistula is not visualized there are usually some characteristic angulations or tenting at the site of the fistula or a telltale streak from the urethra directed posteriorly near the ischial spine. It may document an associated urinary abnormally especially vesico-ureteric reflux. Can demonstrate rare deformities like rectourethral fistula without rectal or anal agenesis and duplication of urethra.[14]

Intravenous pyelography:

It helps to assess the structure and function of the upper urinary tract. Very useful in demonstrating renal dysplasia or agenesis.[14]

***Colostogram*:**

Generally performed before the child goes home or 2 to 3 week after operation or during the early follow up period, around 2 to 3 months of life. This confirms the level of anomaly suspected by invertogram. It may also outline the fistula or may show a 'beak' at the site of fistula. It can give valuable information about the length of the distal loop so that a crucial decision about need for combined abdominal approach can be planned in very high anomalies.[13]

Ultrasound scan of pelvis

Ultrasonography proves to be very valuable in identifying the associated genitourinary abnormalities. The level of the distal pouch can be accurately outlined by careful interpretation of ultrasound. Gas in the distal pouch and full bladder are prerequisite for a proper interpretation. The advantages of ultrasound are simplicity, accuracy, availability in most of the centers now and absence of radiation hazard.[15,16]

Computerized axial tomography (CAT)

It is very useful in the initial assessment of neonates with anorectal anomaly and also can give helpful indications for or against 'Re-do' procedures. CAT can help in demonstrating the site and development of pelvic musculature (sphincter muscle complex). CAT is useful in identifying sacral and spinal abnormality, spina bifida occults, sphincter muscle complex deficiency, (hypoplastic sphincter muscle complex). The conventional axial views best demonstrate the levator sling and its relation to neorectal placement. Coronal views give a better estimate of the bulk of the sphincter muscle complex (SMC). CAT helps to assess the state of SMC and to define the eccentric position of the rectum in postoperative patients with problems.[17]

Magnetic resonance imaging (MRI)

It is the newest diagnostic tool that is capable of generative images of sections of the body in any plane. MRI scanner permits better resolution of soft

tissues than CAT scan. The anatomical relationship of the most distal portion of the bowel to the muscles of continence can be directly visualized with good definition. Impacted meconium in the distal pouch serves as an excellent contrast agent because of high lipid content. The fistula can be accurately identified, lipomas in the sacral spine and other bony abnormalities are well demonstrated and it is believed that they should be corrected before any reconstruction of the rectum (if they are present) so that optimal innervation of muscle will be preserved.

MRI will give important details regarding the length and caliber of the cloacal channel, level of confluence of the urinary, genital and intestinal tracts. MRI is very useful in demonstrative lesions of the spinal cord such as tethered spinal cord or neoplasm, sacral agenesis and thoracolumbar spinal anomalies. Genitourinary tract abnormalities are very clearly outlined by MRI. MRI will be taking an important place in evaluating the level and other abnormalities associated with anorectal malformations in future when the technique is more readily available.[18]

Procedures and Complications

Stephens proposed Sacroperineal or Sacro-Perineo-Abdominal Rectoplasty that through a short sacrococcygeal incision, the plane of the puborectalis is defined by right angled forceps pressing against a metal sound in urethra or vagina; the fistula is ligated and the rectum mobilized either through

the sacral or abdominal incisions, and threaded down through the sling, where it is anastomosed to skin flaps. The essential puborectalis is defined, there is no extrarectal dissection that might interfere with the bladder nerve supply; the anus is skin lined for sensation; and the procedure may be completed by the sacral route alone (for intermediate anomalies) or the abdominal route (if further bowel mobilization is required in high anomalies). The advantages are stresses essential puborectalis, no extrarectal dissection of fistula, permits tapering of bowel, skin lined anus, suitable for reoperations. Disadvantages are puborectalis definitions are 'blind', access to fistula closure limited, does not define external sphincter.

One should not forget, however, that despite these difficulties, in the context of the time of its introduction, Stephens procedure revolutionized the treatment of anorectal anomalies and formed the basis of every advance since then. The results still stand in the forefront of reported series. 68% of patients with high and intermediate anomalies being continent in the series of Stephens and Smith.[19]

Swenson and Donnellan proposed Abdominoperineal Rectoplasty. This procedure is essentially the original Rhoads operation, in which there is no sacral exposure. However, Swenson and Donnellan, appreciating the concepts of Stephens, endeavored to define the correct plane through the puborectalis sling from the abdominal route, and feeding the neorectum through this sling to the perineum (excising any inert or doubtfully ischemic dilated terminal rectum

if necessary). The difficulty is in defining the puborectalis from the abdominal route (where it is hidden behind the bladder and the pelvic fascia), the dissection may involve interference with pelvic parasympathetic outside the rectum and the external sphincter is not defined. The advantages are attempts to define puborectalis, adequate mobilization and tapering, excision of Megarectum. The disadvantages are difficult to define puborectalis, involve pararectal dissection, damage to urethra and nerves and does not define external sphincter. [20]

Stephens – Kiesewette – Rehbein proposed Sacroabdominoperineal Rectoplasty with submucosal Resection, utilizing the concepts of Romualdi avoided any dissection outside the rectum by bringing the neorectum down inside a demucosed sleeve of the original rectum. This step was grafted on to the Stephens sacral approach. Certainly all pararectal tissues are kept intact. Again, however, the puborectal is definition is blind; potential afferent nerve receptors in the rectal mucosa are excised and again little cognizance was given to any potential external sphincter component. Further, the submucosal sleeve dissection necessitates an abdominal laparotomy in every case. Advantages are stresses essential puborectalis, no extrarectal dissection, permits tapering of bowel, accurate identification of fistula and by-pass inert megarectum. The disadvantages are Puborectalis definition is 'blind', sacrifices potential afferents from rectum, and does not define external sphincter.[21]

Mollards Anterior Perineal Rectoplasty approached the sphincter complex by a curved transverse perineal incision anterior to the expected site of

the new anus. A plane to the fistula and to the puborectalis is thus opened up immediately behind the urethra (or vagina), with positive identification of both fistula and levator being visualized. The abdominal portion of the operation is by the submucosal sleeve dissection technique of Rehbein as previously mentioned.[22]

The procedure has the merit of direct identification of the puborectalis, and limited recognition of the external sphincter component, but like the Kiesewetter-Rehbein procedure requires an abdominal mobilization for its performance.[22]

De Vries and Pena' Posterior sagittal Anorectoplasty with a keen awareness of the muscles necessary for control, reintroduced the perineal approach to the rectum. In this the dissection is aided by electrostimulation of all muscle fibers, commencing with precise definition of the maximum confluence of external sphincter components at the proposed anal site. Each muscle is divided in the sagittal plane, including through the combined deep external sphincter puborectalis complex, thus affording a wide access to divide a fistula under vision, mobilize and taper the terminal rectum, and then reconstitute all muscle elements accurately around the neorectum in precisely the correct anatomic position. The exposure is so wide, through an incision from sacrum to anterior perineum that the majority of lesions (even high level anomalies) can be dealt with, without abdominal exposure. There is no doubt that, more than any other exposure, all muscle elements are identified, the bowel

is accurately positioned with respect to these muscles and damage to the urethra is minimized by the wide exposure of the fistula.[23]

The author's criticism, however, relates to the concept of deliberately dividing surgically the entire muscular complex on which eventual continence depends, especially the deep portion (deep external sphincter puborectalis) which is the essential muscle of continence. Further anxieties are the excessive degree of bowel tapering that is advocated and the direct anastomosis of bowel to skin without skin flaps and for both reasons experience is proving that this can result in some stenosis.[23]

Yokoyama proposed Abdomino Extended Sacroperineal Rectoplasty and his colleagues, using electro stimulation and perineal exposure combine some elements of the De Vries – Pena sagittal approach with the Rehbein abdominal submucosal dissection. Although the approach through the perineum does define the puborectalis complex, it does not divide it. The approach to the fistula is via the abdomen, and essentially recognizes a potential internal sphincter in the terminal rectum at the fistula through which the neorectum is tunneled. The external sphincter is identified but not divided. This procedure has the following advantages of no extra rectal dissection, preserves internal sphincter, no disturbance to puborectalis at rectal wall, and utilities external sphincter. Disadvantages of Limited access to fistula, placement through external sphincter.[24]

Perineal Recto plasty:

This procedure is utilized in all intermediate lesions, and (as Pena demonstrates) can be used in many high lesions, such as the standard forms of rectoprostatic urethra fistula and high recto vaginal fistula. Only in the very high lesions or in complex anomalies may it be necessary to open the abdomen.

Perineal Abdomino Perineal Recto plasty:

In some high lesions, insufficient mobilization of the rectum can be achieved by the perineal route. The operation commences in the perineum, with the same exposure as above, and the preservation of the deep sling of the muscle complex, which is defined by a Penrose drain. The patient is then placed in the lithotomy position. The abdomen is then opened and the bowel mobilized; the rectal pouch is preserved and by submucosal sleeve dissection, the fistula is divided from within, the neorectum, tapered if necessary, is brought down within the rectal sleeve to the base of the pouch through which a hole is made to identify the Penrose drain, preserving any circular muscles of the rectal pouch (Hokoyama). From the perineum the tapered bowel is then pulled down through the undamaged sling, and the anoplasty completed by the Nixon technique.[25]

Surgical operations for low anorectal anomalies

Cut back operation

First described by Browne (1951) favoured by most British surgeons, this simple operation is recommended by some authors as The procedure of choice in all low anomalies, especially in females, in the neonatal period. One blade of a pair of straight blunt-tipped scissors is placed in the ectopic bowel orifice, and directed backwards, under the skin, strictly in the midline, to approximately the position of a normal anus. Closing the blades and cutting the intervening tissue between the skin and posterior rectal wall completes the operation. The cut surfaces can be opposed with 4-0 Vicryl or a similar suture, or the raw edges can be left open, as healing is usually rapid. The blade of the scissors must not advance too deeply into the rectum as the encircling fibers of the puborectalis muscle will be divided and subsequent consistence affected.

The cosmetic result with no perineal body is unacceptable to many cultures, and these patients often come back for a secondary transplant of the anus into a more normal position.[26] The functional outcome of the cutback operation should be satisfactory.

Anoplasty:

The advantage, when compared with the repositioning operation, is that the anterior wall remains in close contact with the vaginal and it is not possible to construct a perineal body. Using an electrostimulator, the exact position of

the anus as shown by contraction of the external sphincter can be identified. An inverted ‘V’ or ‘U’ incision is made over the proposed site of the anus and a posterior subcutaneous flap raised, identifying and leaving behind the obvious ‘striated muscle complex’ fibres of the contracting external sphincter. A midline skin incision is made from the opening of the fistula on the perineum to the apex of the inverted ‘V’ incision. The posterior wall of the rectum is defined and cleared of any muscle fibres, cranially, in the median plane, for a distance of about 2 cm from the fistula. When the posterior wall is free, an incision is made, again strictly in the midline, to enlarge the orifice.

The apex of the ‘V’ skin flap is now turned into the gap in the posterior rectal wall and sutured into position with 4-0 Vicryl or similar sutures. The base of the ‘V’ flap must be broad and posterior enough to widen the anal opening to a size of between 10 and 12 mm in diameter. The ‘V’- ‘V’ anoplasty is applicable to both male and female infants with low anorectal anomalies and proves a more satisfactory operation than the simple cutback operation.[27]

Posterior transposition:

This operation is reserved for use in female infants with a low lesion – either an anovesibular or an anocutaneous fistula. The anal transplant operation is not used in the anocutaneous fistula in the male. The operation can be carried out in the newborn period, without a diverting colostomy. It is preferable, from a cosmetic point of view, to the ‘cut-back’ operation.

Position is Supine for this operation. The anal site is identified with a stimulator. A racquet incision is made around the fistula and continued in the midline posteriorly towards the anal site. The incision is deepened into the posterior rectal wall. The identification of the wall of the rectum is aided by a Foley catheter placed in the fistula which inflation of the balloon. By gentle traction on the catheter the rectum can be pulled down, better defined and dissected off the vagina. The opening of fistula is identified and 4-8 traction sutures are placed around the orifice. It is better to err on the side of sacrificing the vaginal wall rather than the rectum, a these vaginal tears will heal spontaneously whereas damage to the rectum may result in a recurrent fistula. Great care should be taken in preserving and dissecting the whole full thickness of the bowel well together with the ‘anus’ or ‘fistula’ as a rudimentary internal sphincter is present at the tip in the normal anatomical position.

The dissection is facilitated if the posterior aspect is freed first leaving the difficult plane of separation between the vagina and rectum to the last. The striated muscle complex is divided only as far as is necessary, the puborectalis is not cut and the rectum is freed only enough to reach the perineum. Too much mobilization may lead to subsequent prolapse of the rectum. Once the rectum is completely freed, the anterior aspect of the wound, between the vagina and the striated muscle, can be repaired with 3-0 vicryl sutures in layers.

A modification is Pott's procedure in which the skin and muscle bridge is maintained and the Rectum rerouted into this. Though, technically difficult it results in a good out come as no structure is divided.

COMPLICATIONS

In a Study by Bliss DP included 355 patients (245 boys and 110 girls) ranging in age from 12-36 months. PSARP was performed in all the patients. There were 195 boys with rectourethral fistula and 95 girls with genitourinary tract fistula while 5 girls had cloacal malformations. Operative and postoperative mortality was 9/355 (2.5%). Early functional results were good in 30%, fair in 45% and poor in 25% patients. Chronic constipation and anal stenosis was found in 99 and 35 patients respectively. Mucosal prolapsed with perineal itching was present in 60 patients. Recurrent UTI and orchitis was found in 5 and 3 patients respectively. Urethral stricture and urethral diverticulum was found in two cases each, while redosurgery was performed in two patients. The incidence of high and intermediate anorectal anomalies was more in male babies. The PSARP procedure is safe with good functional results in terms of faecal continence In the female patients, rectovestibular fistula (46%) and in the males, perineal fistula and imperforate anus without fistula, each with equal frequency (28%) were the most common anomalies. The mean SR in the study group was 0.72 + 0.04. SR was 0.67 + 0.03 among patients who had associated urogenital anomalies, 0.69 + 0.04 in patients suffering from

soiling, and 0.67 +0.02 in patients who had postoperative fecal incontinence. The most common complication following PSARP was soiling (44.9%) and then constipation and fecal incontinence in order of frequency. None of the cases developed urinary incontinence or other urinary complications after PSARP. Seventy-three percent of the patients had voluntary bowel movements (VBMs) and 51% were totally continent. Although the PSARP has a negligible complication rate, the success and outcome of the surgical correction in view of the bowel function depend on the development of the sacral nerves. SR reflects the sacral bone development and can be easily calculated by a pelvic AP film. Considering the lower SR in patients suffering from postoperative soling and fecal incontinence, in comparison to the normal group (0.77), SR could be used as a prognostic index to predict the probability of achieving total continence following PSARP.[28]

A study by Langemeijer RA, there are few follow-up studies comparing posterior sagittal anorectoplasty (PSARP) with conventional procedures for patients with anorectal malformations (ARM). The authors have examined retrospectively postoperative anorectal function of patients with ARM treated with PSARP compared with those treated with conventional methods. Anorectal function in 23 patients with high and intermediate type anorectal malformations (PSARP group), who underwent PSARP more than 4 years previously, were assessed by Kelly's clinical scoring system and objective studies. These results were compared with those in 14 cases (5 high and 9

intermediate type cases; control group), who underwent other conventional surgical procedures. Results: Using Kelly's clinical scoring system, scores of the PSARP group compared with the control group were good in 48% versus 21%, fair in 48% versus 58%, and poor in 4% versus 21%, respectively. Barium enema studies suggested better anorectal sphincteric function in patients with high anorectal malformation in the PSARP group. Magnetic resonance imaging (MRI) studies showed more correct placement of the rectum through the striated muscle complex in the PSARP group at the I-line level. Manometric studies showed no difference in maximum resting pressure, anal canal length, and the incidence of anorectal reflex between the two groups. The favorable results of MRI and barium enema studies can be explained by direct visualization of the striated muscle complex with the aid of electrical stimulation as well as no harmful effects of amputation of the sphincter muscle in PSARP. However, manometric studies suggest anorectal function in patients with high and intermediate anorectal malformations is limited even after PSARP. Long-term postoperative follow-up with adequate bowel management is required for all patients with high or intermediate anorectal malformation.[29]

Aims and objectives

- To study the Epidemiology of Anorectal Malformation.
- A demographic survey of the presenting cases.
- Follow up of the cases and their outcome.
- To study different surgical procedures to correct Anorectal Malformations with regards to it complications.
- Analysis of the cases which did not do well.

Patients and methods

INCLUSION CRITERIA

All cases of ARM admitted in GRH, Madurai during the period from September 2008 to March2011, completed all the stages of surgical procedures with in this period and with the follow up of 6 months.

EXCLUSION CRITERIA

Nil

STUDY PERIOD

September 2008 to March 2011.

PLACE OF STUDY CONDUCTED

Dept. of Paediatric Surgery, Govt. Rajaji Hospital, Madurai.

METHODOLOGY

All babies with ARM patients admitted to GRH are first stabilized. In newborn male child, child is kept under observation for16 to 24 hours. The child is subjected to through clinical examination. If there is clinical evidence of perineal fistula, bucket handle deformity or mid line raphe fistula the child is subjected to Anoplasty. Child is followed with anal dilatation. If in first 24 hours if there is meconuria and flat bottom the child is subjected to colostomy. Then at 6 months child is taken for definitive procedure (PSARP) done. If no clinical evidence of type of anomaly then invertogram is taken at 16 to 24 hours.

If on invertogram low ARM is diagnosed child is subjected to Anoplasty. If high or intermediate anomaly, child is subjected to colostomy and followed at 6 months with definitive procedure. In all cases Colostogram is taken before definitive procedure. After 3 months colostomy closure was done. In newborn female child, child is kept under observation for16 to 24 hours. The child is subjected to through clinical examination. If there is clinical evidence of cutaneous fistula, the child is subjected to Anoplasty. Child is followed with anal dilatation. In case of anovestibular fistula or vulvar anus/fistula posterior transposition is done at 6 months and followed with anal dilatation. In case of cloacal anomaly, colostomy is done, followed by definitive procedure (PSARVUP) at 6 months. If no clinical evidence of fistula then invertogram is taken at 16 to 24 hours. If on invertogram low ARM is diagnosed child is subjected to Anoplasty. If high or intermediate anomaly, child is subjected to colostomy and followed at 6 months with definitive procedure. In all cases associated anomalies are diagnosed during first month of life by subjecting to investigations.

OUTCOMES MEASURED

1. Mortality
2. Morbidity and Complications

Results

Table - 1 : Socio-economic status

Income	<RS12000/Annum	> RS12000/Annum
Number of patients	90	0

In our study all patients are in a < 12000/Annum income group, no patients belong to > 12000/Annum income group. All the patients where in our study were low socio-economic status.

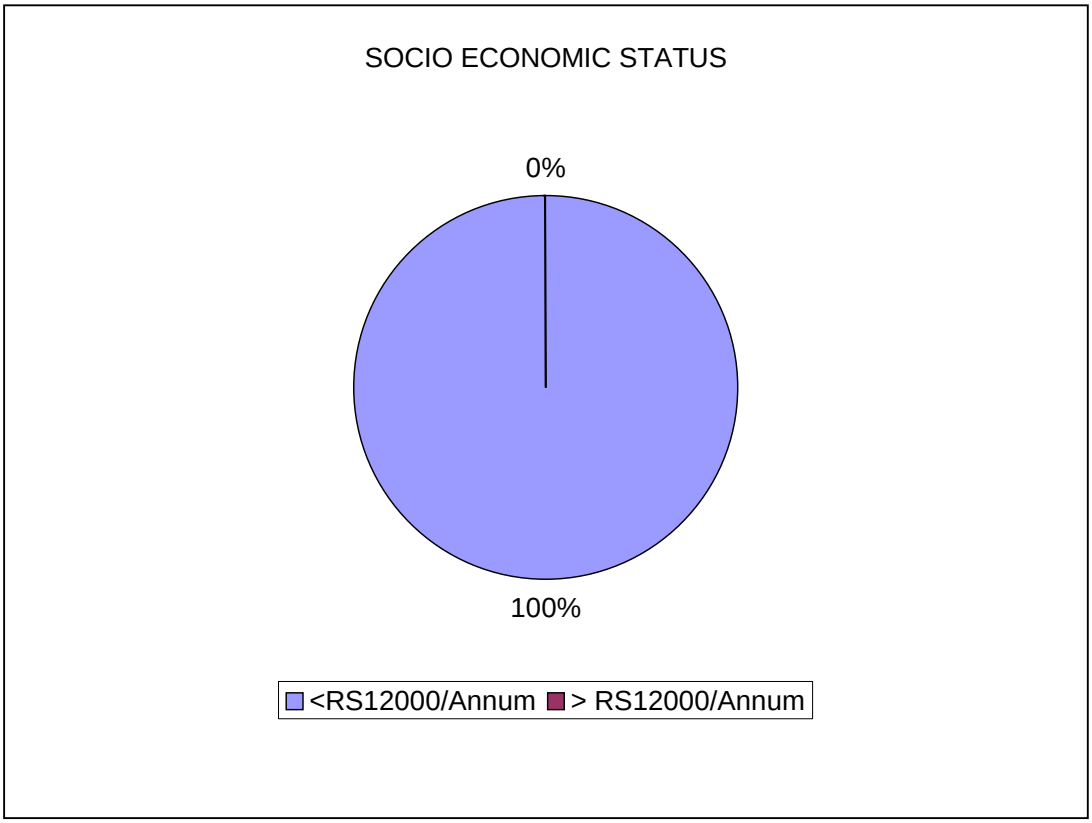


Table - 2 : Sex Ratio

Gender	Our study	Associated anomalies
Male	55%	67%
Female	45%	33%

In our study, the male: female ratio associated with ARM is almost equal, with a 55:45 male: female ratio. In the same study, males were said to have a 2:1 incidence of associated anomalies than girls.

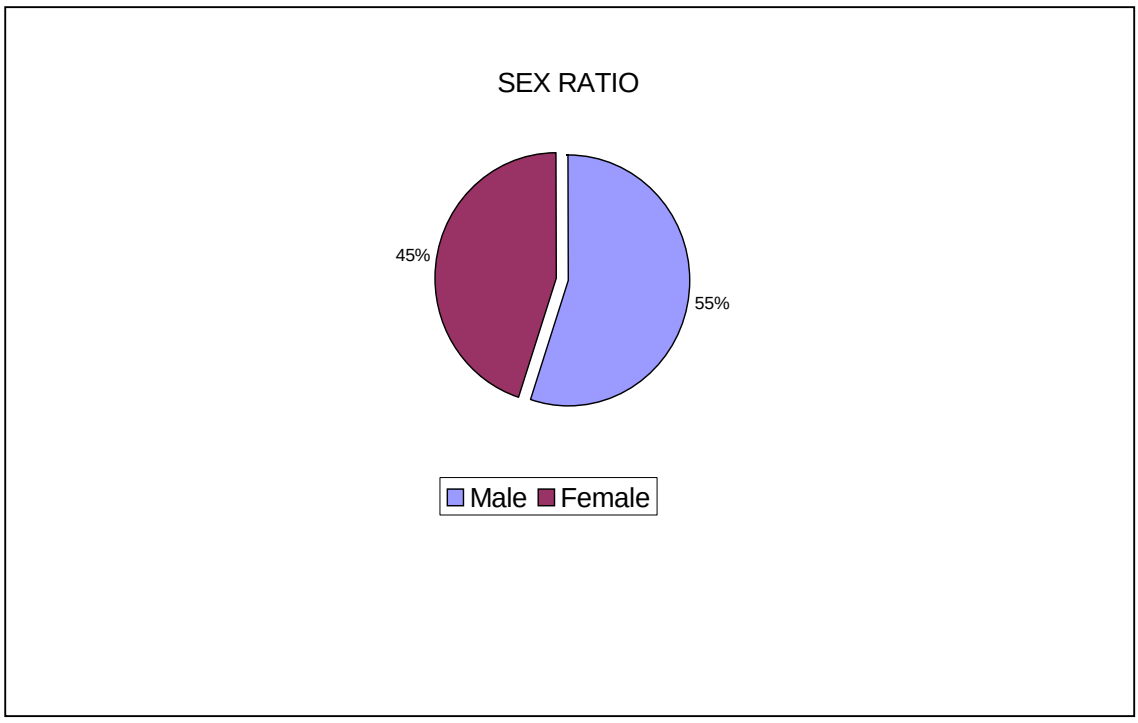


Table – 3 : Types of Fistula in ARM

No.	Type of fistula	Our study	
		Male %	Female %
1	Rectourethral fistula	30	
2	Rectocloacal fistula		5
3.	Recto Vesical fistula	10	2
4.	Recto vaginal fistula		15
5.	ARM – No Fistula	2	2
6.	Anterior Perineal anus		12
7.	Anovestibular Fistula		29
8.	Vulvar Anus		7
9.	Complete covered Anus	10	7
10	Anocutaneous fistula	28	12
11.	Rectal Atresia	2	2
12.	Pouch colon	2	
13.	Rectovestibular fistula		5

In our study of 90 cases, most common type of fistula in male is rectourethral fistula (30%) and in females it is anovestibular fistula (29%). Cloacal anomaly (5%), Recto vaginal (15) and malpossitioned anus – Anterior perineal anus (12%), Anovestibular fistula (29%), Recto vestibular fistula (5%) and Vulvar anus (7%) found exclusively in females.ARM without fistula are found equally in males and females (2%).

Rectovesical fistula found more in males (10%) than in females (2%). Rectal atresia which is common in south India is found in equal distribution in male and female (2%).

Types of Fistula in ARM

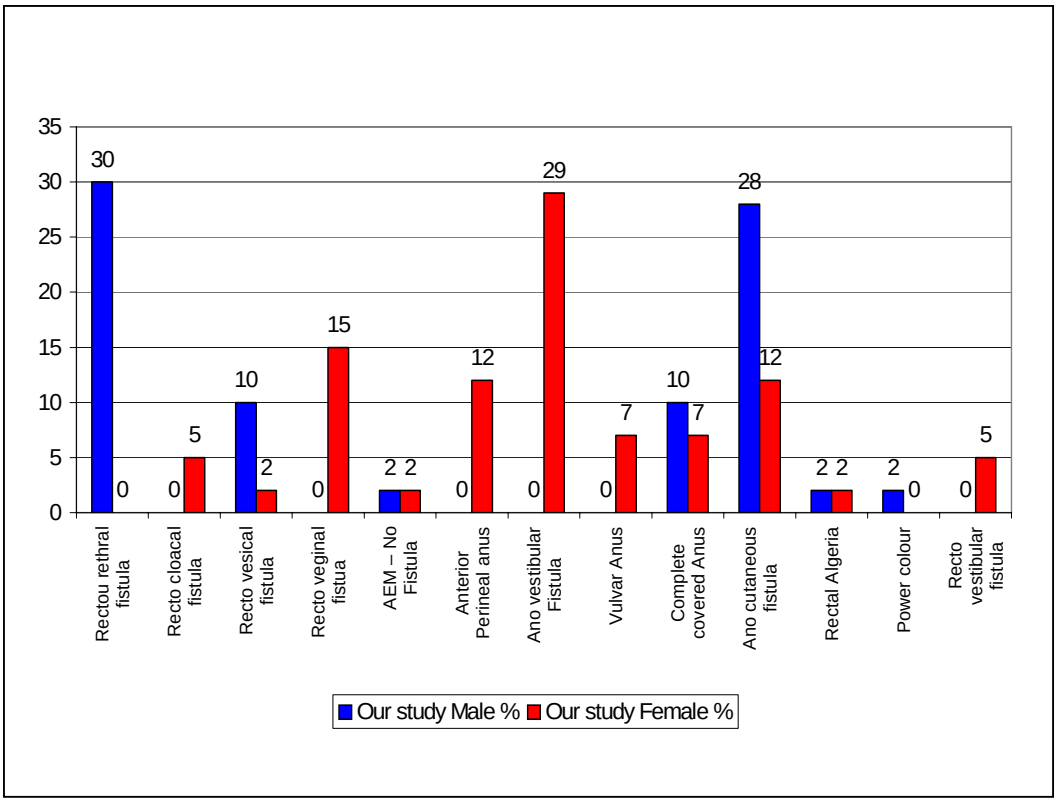


Table – 4 : Anomalies Associated with Anorectal malformation

Type of anomalies	Our study
Anomalies %	48
Vertebral / Skeletal	44
Cardio vascular	23
EA / TEF	7
GIT	7
GU	35
Genitalia	9
Downs	2

In our study of 90 patients , 48%(44) patients has associated anomalies out of which most common association is Vertebral/ skeletal anomalies(44%) followed by Urological anomalies(35%), Cardiovascular(23%), Genitalia(9%), GIT(7%), EA/TEF(7%) and Downs syndrome(2%)

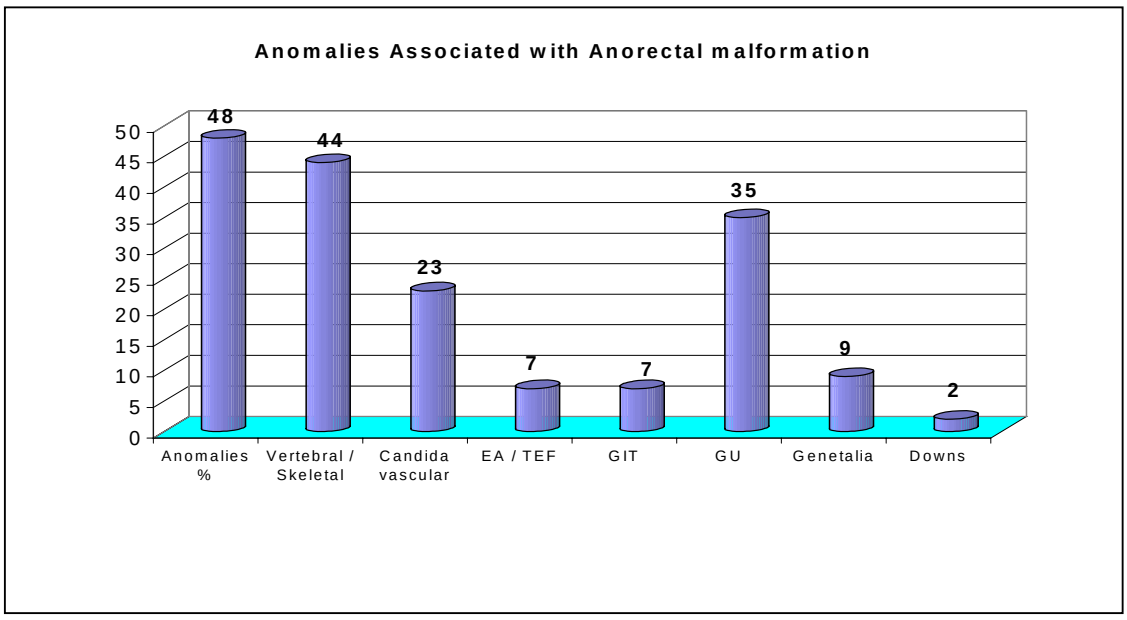


Table – 5 : Type of Anomaly

	Male	Female	Total
High	17	10	27
Intermediate	12	4	16
Low	19	28	47
Total	48	42	90

In our study most common anomaly in female is Low anomaly(28) and in males is High (High and Intermediate) - 28 cases

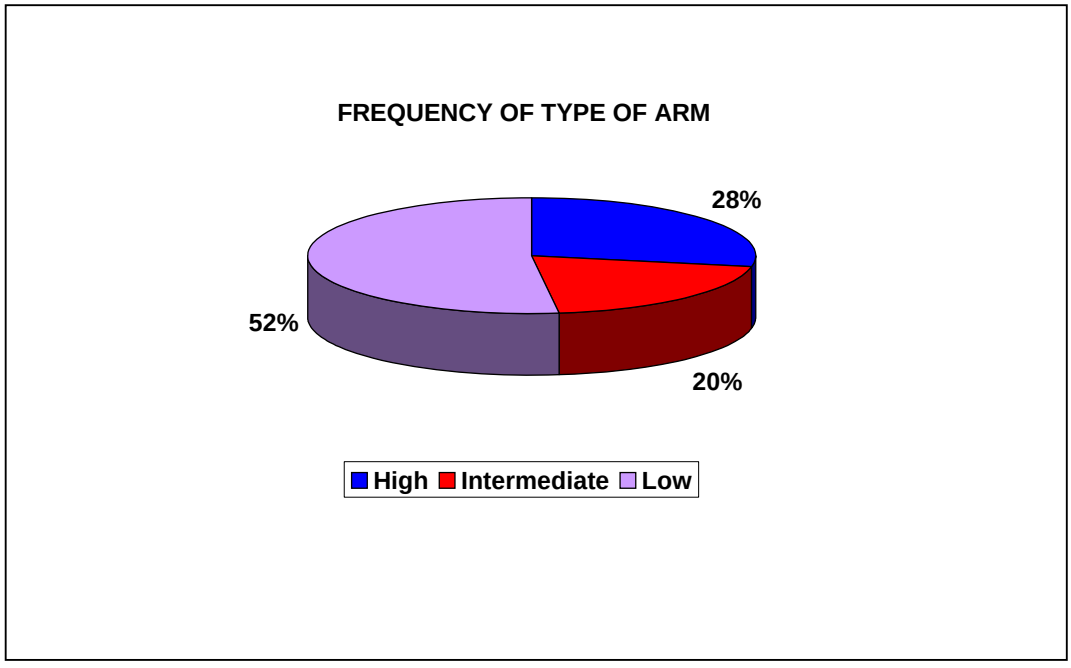


Table - 6 : Correlation between invertogram and operative finding

	Invertogram	Operative findings
High	23	18
Intermediate	11	12
Low	4	8

In our study out of 23 cases diagnosed as high by invertogram 18 were confirmed to be high. In the same way11 cases diagnosed as intermediate by invertogram 12 were confirmed to be intermediate by operative finding and 4 cases diagnosed as low by invertogram 8 were confirmed to be low by operative finding

Table – 7 : Complications of colostomy

Complications	Our study
Skin excoriation	21
Prolapse	14
Bleeding	12
Obstruction	5
Wound infection	2
Retraction	5
Revision Rate	5
Mortality	5

In our study 39 colostomies was done out of which 23 were pelvic colostomy and 16 were transverse colostomy. 21% patients had skin excoriation, 14% had prolapsed of colostomy, 12 % had bleeding from colostomy, 5 % had intestinal obstruction and 2% had wound infection.

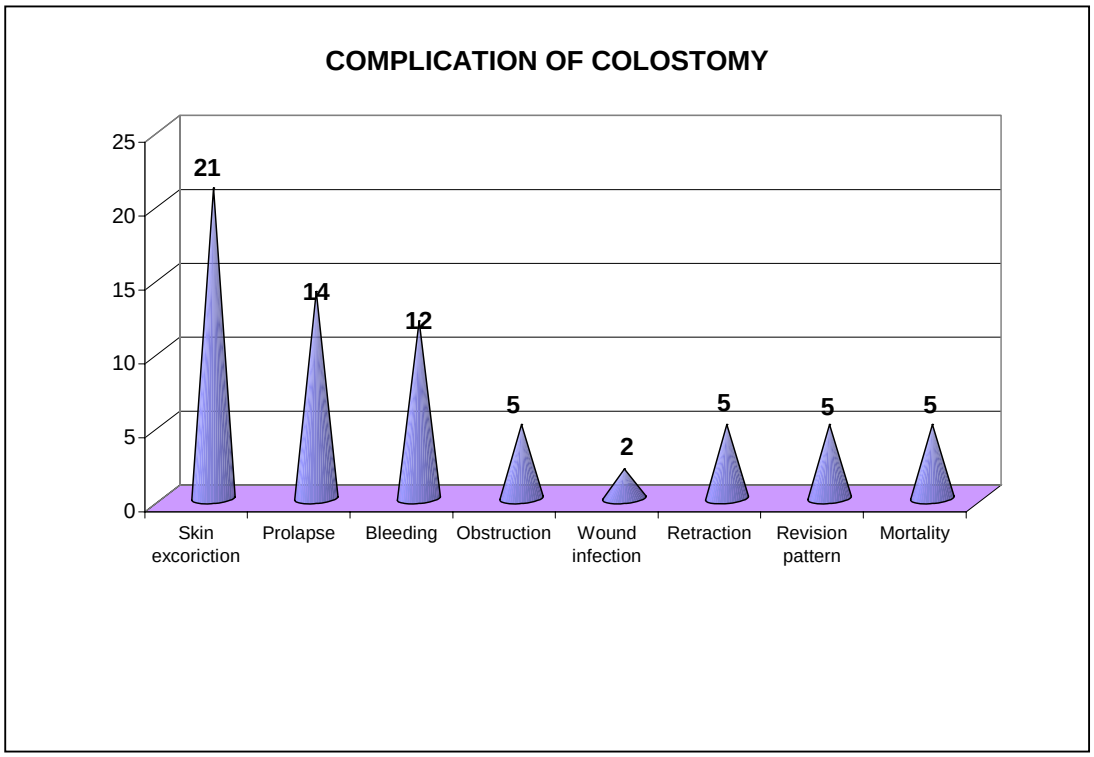


Table – 8 : Complication of colostomy closure

	Complication rate
Wound infection	15.6
Anastomotic leak	6.2
Adhesive obstruction	6.2
Stitch granuloma	12.4
Incisional hernia	3.1
Mortality	-

Wound Infection is the most common complication of colostomy closure(15.6) followed by Stitch granuloma(12.4), Anastomotic leak and Adhesive obstruction(6.2) and Incisional hernia(3.1)

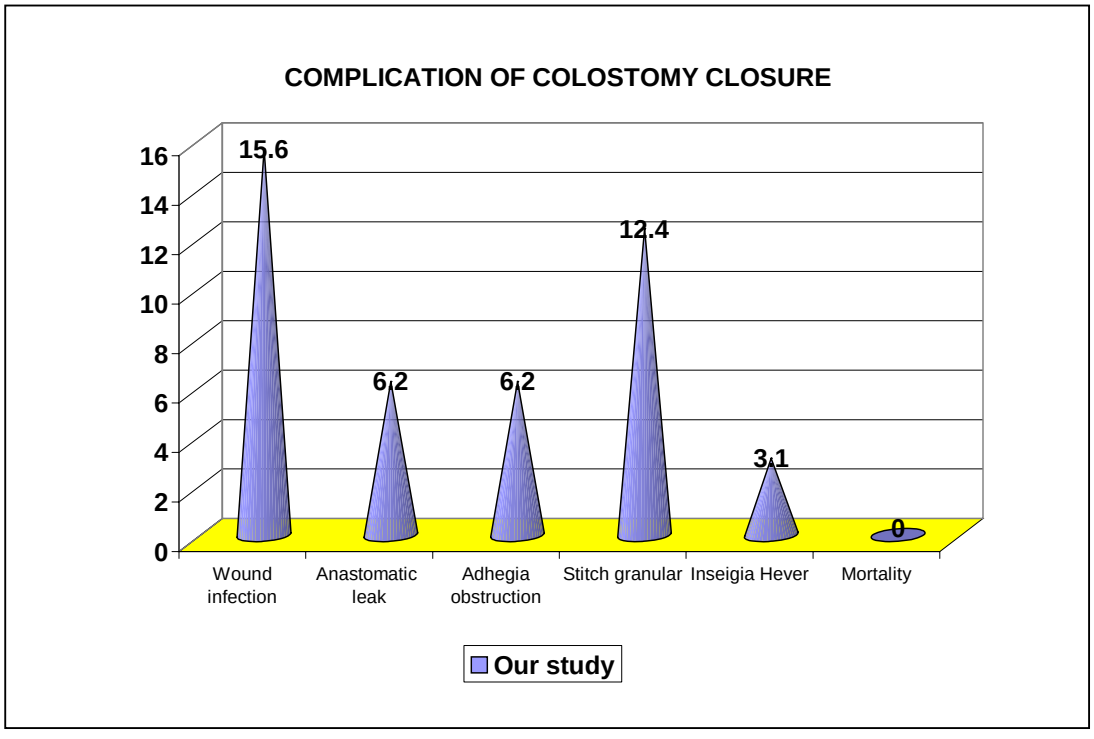


Table – 9 : Complications of posterior sagittal approach & Lap. Assisted approach

	Post sagittal approach	LAAPT
Wound sepsis	0	1
Wound dehiscence	2	0
Retractions	2	1
Faecal fistula	1	0
Rectal stricture	0	1
Anal stenosis	1	2
Mucosal prolapsed	3	1
Redo Anorectoplasty	3	1
Redo Anoplasty	1	2
Laparotomy needed	11	
Continence		
Good	45	48
Fair	33	32
Poor	22	20

A comparison of laparoscopic assisted (LAARP) and posterior sagittal (PSARD) anorectoplasty in the outcome of intermediate and high anorectal malformation, There is no significant difference in complication rate between two procedures

Table – 10 : Low Anomalies

Type of procedure	Number of cases in our study	Complications	
		Stricture	Redo
Anoplasty	27	2	1
ASARP	7		
Posterior transposition	12	1	1

In our study totally 3 procedures are done for Low Anorectal Malformations. We have done Anoplasty in 27 patients, ASARP in 7 patients, and Posterior transposition in 12 patients. Out of 2 Anoplasty which went in for stricture, Redo Anoplasty was done in 1 patient. 1 Posterior transposition went in for stricture which was subjected to redo Posterior transposition.

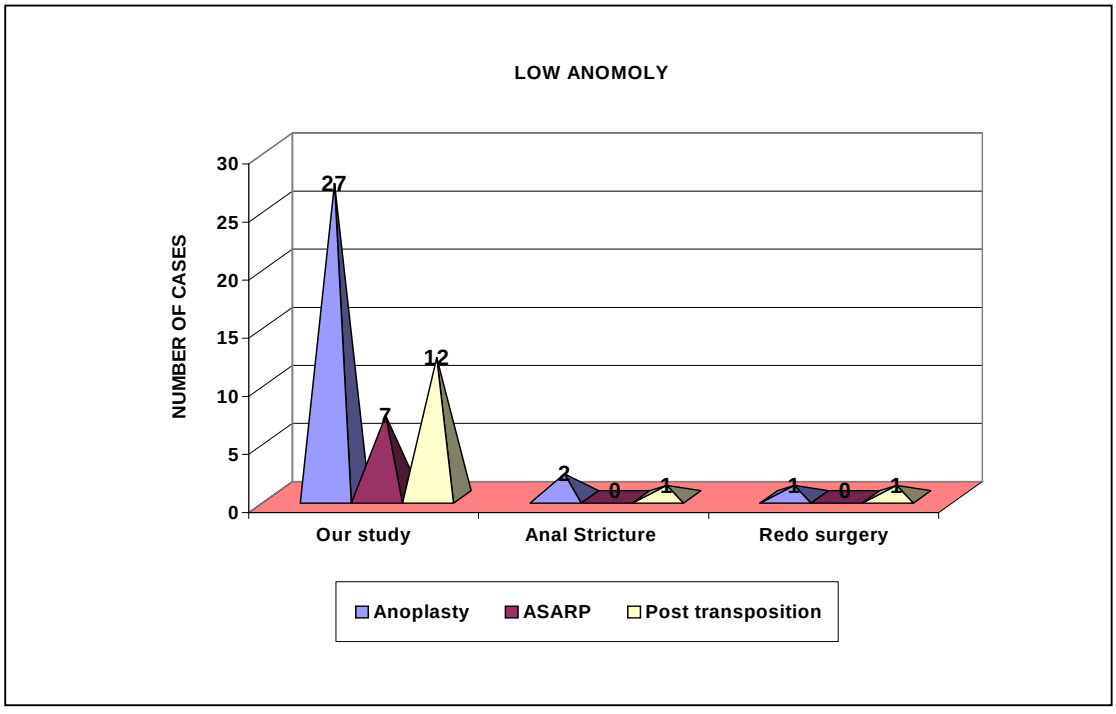


Table – 11 : Functional Outcome of High and low Ano rectal malformations

Kelly Score	Good	Fair	Poor
High ARM	47	33	20
Low ARM	85	13	2

Functional outcome of Low ARM in our study was Good in 85, Fair in 13, Poor in 20 patients and for High ARM it is Good in 47, Fair in 33 and Poor in 20 patients.

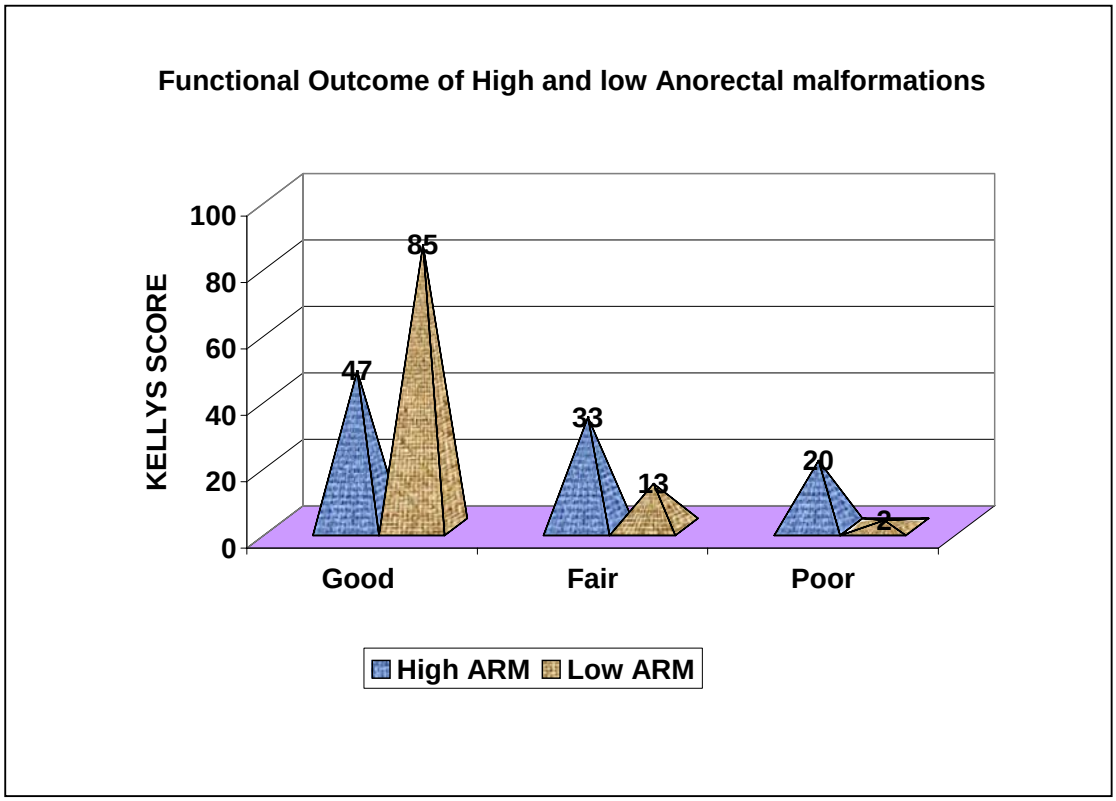
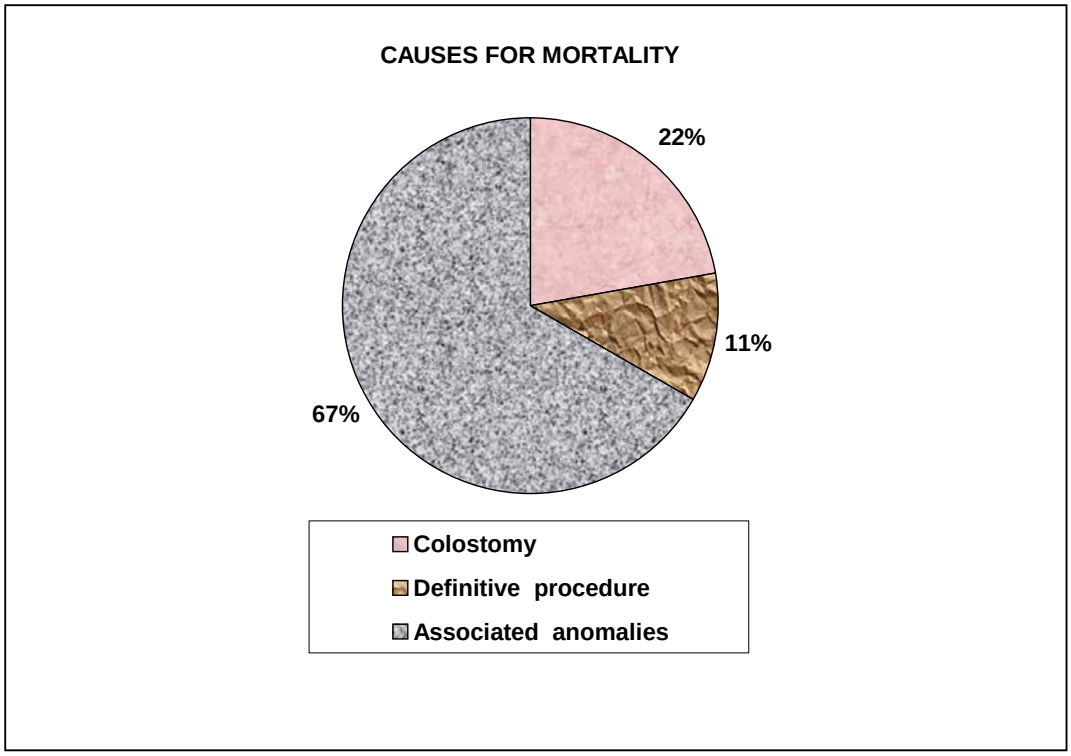


Table – 12 : Mortality

Cause for Mortality	Number of cases
Colostomy	2
Definitive procedure	1
Associated anomalies	6
Total	9

Total mortality in our study is 9. Two patients due to colostomy,1 due to definitive procedure and 6 due to associated anomalies



Discussion

Table - 1 : Socio-economic status

Income Group	Our study		A. E. Archibong, et al.[1]	
	Less than Rs. 12000/annum	*Mores than Rs. 12000/annum*	*Less than Rs. 12000/annum*	*More than Rs. 12000/annum*
Number of patients	90	_	125	30

The incidence of ARM in affluent patients with income group >1200 is very meager and practically nil. I understand from my senior colleagues who practice outside that ARM predominantly affects lower socioeconomic status patients. According to A. E. Archibong low socio economic group patients who were exposed to various harm full agents such as poverty, ignorant are also affected with higher incidence of ARM. In his study also ARM is very low in affluent patients.

Table - 2 : Incidence

	1ST YEAR	2ND YEAR	3RD YEAR
Number of Cases of ARM	36	30	24

The incidence of ARM is decreasing. Previously in the yester years we were having a heavy workload because of these congenital diseases. Now, the number of cases has come down. Reasons probably may be due to the better nutritional status and better ante natal care which our people enjoy. Further the Ultra Sound may help in discerning there afflicted babies leading to spurning of such fetuses which suffer from malformation disorders. This is reflected in this study as evinced by the decreasing incidence noted through the years.

Table – 3 : Sex Ratio

Gender	Stephens[30]	Endo et al[31]	Our study
Male	57%	57%	55%
Female	43%	43%	45%

In our study, the male : female ratio associated with ARM is almost equal, with a 55 : 45 male : female ratio. Our results are almost similar to the study of Stephens and Endo et al. In these study also male female ratio is almost equal. There is no difference in incidence of ARM in both the sex group even though certain type of ARM is exclusive for either male or female gender.

Table – 4 : Types of Fistula in ARM

No	Types of fistula	Our study		Stephens & Smith[19]	
		Male %	Female %	Male %	Female %
1	Rectourethral fistula	30		36	
2	Rectocloacal fistula		5		5
3.	Rectovesical fistula	10	2	6	5
4.	Rectovaginal fistula		15		19
5.	ARM – No Fistula	2	2	8	4
6.	Anterior Perineal anus		12	4	17
7.	Anovestibular Fistula		29		18
8.	Vulvar Anus		7		
9.	Complete covered Anus	10	7	10	4
10	Anocutaneous fistula	28	12	25	18
11.	Rectal Atresia	2	2		
12.	Pouch colon	2			
13.	Rectovestibular fistula		3		

Though ARM is distributed equally in both the sex, in concurrence with study by Stephen and Smith Anocutaneous fistula has male preponderance and ectopically placed anus has female preponderance. Certain types of ARM such as Cloacal anomaly, recto vaginal fistula and Anovestibular fistula are exclusive for females. In our study there are 2 cases of rectal atresia 1 in male and other in female, which is not seen in study by Stephen and Smith. Rectal atresia is a rare anomaly according to Stephen and Smith but its incidence is high in Southern parts of India as stated by Prof. T. Dorairajan. Pouch colon is other anomaly which has high incidence in Northern India but not in south India, as

stated by Prod. Wakhlu. This shows Geographic variation in incidence of ARM.

Table – 5 : Frequency of type of ARM

Authors	High %	Intermediate%	Low %
Our study	28	20	52
Cook[32]	28	23	51
Stephens[19]	46		54
Chen[33]	20	47	33
Endo et al[31]	26	11	57

In our study Low anomaly is most common occurring anomaly followed by High and Intermediate anomaly, which is also shown by other studies – Cook et al and Endo et al. Difference in incidence shown by Chen et al and Stephen et al is because of difference in classification of ARM used by them. We used International Classification of ARM.

Table – 6 : Anomalies Associated with Anorectal malformation

	Our study	Ratam[34]	Smith[25]	Kiesewetter[35]
Anomalies %	48	58	61	54
Vertebral / Skeletal	44	41	26	6
Cardio vascular	23	10	9	7
EA / TEF	7	6	4	9
GIT	7	9	8	4
GU	35	39	25	40
Genitalia	9			
Downs	2			

Almost half of the cases with ARM has associated anomalies (48%) and it is the most common cause of death in ARM patients (6 out of 9 mortalities in our study). It is recommended that all patients with anorectal malformations should have all necessary investigations to search the associated anomalies different systems. Urinary anomalies were high in both sexes in high ARM. Patients with urogenital anomalies require careful assessment and timely intervention for better out come.

However large number of patients and poor primary health care services make us confine to do basic investigations rather than follow a protocol. So we routinely do Ultrasonography of abdomen, X-ray spine of all patients, echocardiogram and neurosonogram. Special investigations for example: intravenous Urography, MCUG and fistulogram are done in appropriated cases.

Actual incidence of urogenital anomalies may be higher if thoroughly investigated.

Table - 7 : Correlation between invertogram and operative finding

	Invertogram	Operative findings
High	23	18
Intermediate	11	12
Low	4	8
Total	38	38

.

The overall sensitivity of invertogram in detecting type of anomaly is low. In our study out of 23 cases diagnosed as high by invertogram 18 were confirmed to be high, four were low and one intermediate, totaling five. In these five cases, four of them would have been subjected to colostomy because of the wrong vagaries of the Invertogram. So, we would like to stress, that though Invertogram is being done as a routine, clinical assessment is the ultimate parameter for judgment. Cases with epithelisal pearls, bucket handle deformity are pathognomic of low anomaly and are treated with perineal exploration irrespective of Invertogram findings. Surasak Sangkhathat

suggested high sensitivity and specificity with MRI. So through clinical examination is needed if possible MRI for diagnosis of type of ARM

(e.g. ; avoids 3 staged procedure for misdiagnosed Low type ARM).

Table – 8 : Comparison between Pelvic and Transverse colostomy

No.		Pelvic colostomy	Transverse colostomy
		%	%
1.	Skin excoriation	17.3	31
2.	Prolapse	13	18.7
3.	Bleeding	8.6	18.7
4.	Obstruction	-	12.5
5.	Wound infection	4.3	-
6.	Retraction	4.3	6.2
7.	Redo	4.3	6.2
8.	Mortality	-	12.5

The common complication of colostomy is skin excoriation 17.3% in pelvic colostomy and 31 % in transverse colostomy. Similarly all the other complications such as Prolapse, Bleeding, Obstruction, Wound Infection, Retraction, and Mortality are more in with Transverse colostomy than with Pelvic colostomy. In addition to the above complications Transverse colostomy have additional complications such as electrolyte imbalance and malnourishment problems. This show that Pelvic colostomy is better option

when compared to Transverse colostomy which was also in accordance with study by Chandramouli .Sigmoid colostomy should be performed whenever possible except in situations of very high anomaly where surgeon suspects that distal bowel won't be sufficient for further pull through procedures. Close attention to technical details, principles of stomal care, and proper parental instruction should minimize morbidity. Concluding that while transverse colostomy is surgeon friendly for subsequent procedures, pelvic colostomy is patient friendly for maintenance and lesser complication rate.

Table – 9 : Complication of colostomy closure

	Our study	Chandraemouli[36]
Wound infection	15.6	12.6
Anastomotic leak	6.2	7.1
Adhesive obstruction	6.2	5.2
Stitch granuloma	12.4	10.5
Incisional hernia	3.1	2.6
Mortality	-	1.8

In our study wound infection occurred in 5 cases(15.6%). Incisional hernia in 1 case in which wound infection was very severe. Anastomotic leak occurred in 2 cases and were managed conservatively. Adhesive obstructions in 2 cases were also managed conservatively. All this showed that early colostomy

closure reduces morbidity and mortality of colostomy. Hence, it is recommended that Post PSARP patients should have their colostomy closure within three months to obviate complications of the pulled through bowel. Frequently we see patients coming years after the primary procedure. On our part we should motivate and give dates with in three months for the patients at the time of discharge after PSARP.

Table – 10 : Complications of posterior sagittal & Lap. Assisted approaches

	Our study		C. Devos, M. Arnold et al[37]	
Complications	Post sagittal approach	LAAPT	Post sagittal approach	LAAPT
Wound sepsis	0	1	0	2
Wound dehiscence	2	0	2	0
Retractions	2	1	1	1
Faecal fistula	1	0	1	0
Rectal stricture	0	1	0	1
Anal stenosis	1	2	1	3
Mucosal prolapse	3	1	3	2
Redo Anorectoplasty	3	1	3	0
Redo Anoplasty	1	2	1	0
Laparotomy needed	11			
Continence				
Good	45	48	48	43
Fair	33	32	30	30
Poor	22	20	30	25

A comparison of laparoscopic assisted (LAARP) and posterior sagittal (PSARP) anorectoplasty in the outcome of intermediate and high anorectal malformation showed no significant difference in complications of

assisted (LAARP) and posterior sagittal (PSARP) anorectoplasty but have specific associated problems. The increased association of anal stenosis in the LAARP procedure might be due to the fact that the perineum is not as extensively opened as in PSARP, leading to a smaller fashioned anoplasty. However, PSARP group showed a high number of patients needing management for both prolapse. Although a long 'learning curve', with laparoscopic surgical techniques, extending to all participating staff and even equipment maintenance. Both the LAARP and PSARP procedures can successfully treat ARM with comparable outcomes. It appears that LAARP is optimal for high ARMs that would otherwise require a laparotomy to facilitate adequate mobilization.

We suggest that were sacro abdominal pull through is contemplated a lap assisted PSARP would be of value as it obviates the need for laparotomy and it is not a very technically demanding procedure through the laparoscope.

Table – 11 : Complications of posterior sagittal approach & Lap. Assisted approach

	Post sagittal approach	LAAPT	Chi square value	
Wound sepsis	0	1	0.663	Not Significant
Wound dehiscence	2	0	0.961	Not Significant
Retractions	2	1	0.663	Not Significant
Faecal fistula	1	0	0.604	Not Significant
Rectal stricture	0	1	0.663	Not Significant
Anal stenosis	1	2	0.469	Not Significant
Mucosal prolapsed	3	1	0.645	Not Significant
Redo Anorectoplasty	3	1	0.645	Not Significant
Redo Anoplasty	1	2	0.469	Not Significant
Laparotomy needed	11	0	0.122	Not Significant
Continence				
Good	45	48		
Fair	33	32		
Poor	22	20		

Table – 12 : Functional Outcome of High Ano rectal malformations

Kelly Score	Good	Fair	Poor
Our Study	47	33	20
Stephen and Smith[19]	56	32	12
Trustler & Willkinson[38]	26	20	54
Partridge and Gough[39]	33	43	24
Taylor[40]	24	20	56

The continence scoring of our procedure (Posterior sagittal approach) for high and intermediate ARM is better compared to other traditional procedures studied in by different authors Trustler & Willkinson, Partridge and Gough and Taylor.

Table – 13 : Functional Outcome of Low Ano rectal malformations

Kelly Score	Good	Fair	Poor
Our Study	85	13	2
Stephen and Smith[19]	83	15	2
Trustler & Willkinson[38]	80	20	-
Partridge and Gough[39]	86	11	3

The functional outcome patients treated with low ARM is good. This is also substantiated by other studies by Stephen and smith, Partridge and Gough, Trusteler and Willkinson. This is due to less complexity of the defect and good sphincter muscle complex development. 5 patients had fair out come out of which 4 did not turned up for regular dilatation. 1` patient had anal stenosis due to ischemia for which redo surgery was done with poor out come. A typical problem in treatment of low anomaly is anal stenosis which can be prevented by regular dilatations.

Table - 14 : Mortality

Causes	Number of patients
Colostomy	2
Defining procedure	1
Associated Anomalies	6
Total	9

The most common cause of death in ARM patients is Associated Anomalies. Severe forms of anomalies are associated more often with high ARM). It is recommended that all patients with anorectal malformations should have all necessary investigations to search the associated anomalies different systems. Next common cause of death in our study is due to colostomy. These cases presented very late and had a morbid pre-operative picture itself. Early colostomy closure reduces morbidity and mortality of colostomy

Summary

I have studied the present series of 90 patients who were admitted with ARM and underwent various surgical procedures. Analyzing them, the following summary were drawn

1) All the patients where in our study were low socio-economic status. The incidence of ARM in affluent patients with income group >1200 is very meager and practically nil

2) The male: female ratio associated with ARM is almost equal, with a 55:45 male: female ratio.

3) Low anomaly is common in females and high and intermediate anomaly in males. Geographic variation in incidence of ARM in case of rectal atresia and pouch colon. Rectal atresia which Madurai has a higher incidence is also reflected in this study.

4) Almost half of the cases with ARM has associated anomalies (48%) and it is the most common cause of death in ARM patients (6 out of 9 mortalities in our study). which most common association is Vertebral/ skeletal anomalies(44%) followed by Urological anomalies(35%). It is these associations which decide the prognosis rather than the lines of management.

5) The overall sensitivity of invertogram in detecting type of anomaly is low. Invertogram is done as a matter of routine but should not be taken as a fool proof investigation. Clinical determinants are the deciding norms.

6) Pelvic colostomy is better option when compared to Transverse colostomy. Sigmoid colostomy should be performed whenever possible except in situations of very high anomaly where surgeon suspect that distal bowel would not be sufficient for further pull through procedures.

7) Early colostomy closure reduces complications. Frequently we see the proximal bowel is dilated and the distal loop has a very small lumen which results to a anastomosis like end on back, as done in Atresias. This is due to disuse atrophy of the distal bowel. Hence a plea is made for early closure of the colostomy after PSARP.

8) Both the LAARP and PSARP procedures can successfully treat ARM with comparable outcomes. It appears that LAARP is optimal for high ARMs that would otherwise require a laparotomy to facilitate adequate mobilization.

9) The functional results of posterior sagittal approach is better than traditional procedures for High and Intermediate anomalies.

10) The functional outcome patients treated with low ARM is excellent.

11) The most common cause of death in ARM patients is Associated Anomalies.

Conclusion

The outcome of treatment of ARM has improved, tremendously in the present scenario. What was previously a write off are now becoming correctable and these patients lead a normal life, enter adult hood, and bear children. This is because of improvement in the infra structure, availability of adequate health resources, improvement in techniques and investigation modalities. Further, understanding the complex anatomy of ARM and associated anomalies and early and appropriate surgical treatment, emphatically reduces the mortality and morbidity. The Posterior Sagittal Approach has improved functional outcome of treatment of high and intermediate ARM.

The really bad cases are also being discerned by Ante natal Ultra Sound and eased off from the picture. Surgical procedures have become standardized and PSARP has become the state of the art procedure. Laparoscopy has become a tool which also helps in the high Supra Levator anomalies, giving equally good if not better results. Complex Cloacal anomalies are now correctable technically, which was previously not possible.

Gross defects with associated anomalies, are bearing the crux of the mortality. Though ARM can be corrected the other anomalies do the patient in. Corrections are now possible at an earlier date, and the patients are fully corrected before the school going age. Technically continuity of the Gastro

intestinal tract can be restored irrespective of the level of the lesion but the issue on in continence is yet to addressed.

Further, Paediatric Surgery is a service field and may not offer financial overtures. Hence, the Government must announce ARM as a disability and offer the patients the assistance extended to the disabled.

All said and done prevention is better than cure and ARM should be reduced if not preventable. Patient education on Nutritious food, Folic and Iron acid supplementation should be emphasized, and the Government can give concessions to the women in the Antenatal period.

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Abbreviations (in Master Chart)

M	-	Male
F	-	Female
PC	-	Pelvic Colostomy
TC	-	Tranverse Colostomy
Rt	-	Right
Lt	-	Left
VUR	-	Vesico Ureteric Reflex
OA & TEF	-	Oesophageal atresia and Tracheo Esophageal fistula
VSD	-	Ventricular Septal Defect
ASD	-	Atrial Septal Defect
PDA	-	Patent Ductus arteriosus
TOF	-	Tetrology of Fallot
HN	-	Hydronephrosis
HPS	-	Hypospadias
AV	-	Atrio Ventricular
PSARP	-	Posterior Sagittal Ano Recto plasty
ASARP	-	Anterior sagittal Ano Recto Plasty
Post.	-	Posterior
Obst	-	Obstruction
WI	-	Wound Infection
LAAPT	-	Laparoscopic Assisted Pull Through
PT	-	Pullthrough

MASTER CHART

S.No.	IP No.	Name	Age	Sex	Weight	Type of Fistula	Associated Anomalies	Ostomy	Procedures	Socio-economic status	Complications			Mortality	Kelly Scoring continence	Follow up
											Ostomy	Definitive procedures	Colostomy closure			
1	70953	Keerthana	7	F	10	Ano Vestibular fistula			ASARP	low					Fair	+
2	16087	Chandran	6/12	M		Recto bulbar fistula	Patent Urachus	PC	LAAPT	low		Retraction, Ano rectoplasty			Poor	+
3	73879	Roja	8	F	15	Ano Vestibular fistula			Anoplasty	low					Fair	+
4	77003	Sridhar	1	M	8.5	Recto bulbar fistula	Bifid scrota	PC	PSARP	low	Skin erosion	wound dehesion			Poor	+
5	69467	B/o Selvi	1/365	M	2	ARM - High	Complex cardiac defect	TC		low				Death		+
6	89205	Rupika	10/12	F	8	Recto vaginal fistula (Low)	Lt Uretrocele	TC	PSARP	low	Prolapse bleeding		WI, Incis. hernia		Fair	+
7	94055	Pon Sridhar	1	M	9	Recto prostatic fistula	Hemi vertebra	PC	LAAPT	low		Anal stenosis, wound sepsis			Fair	+
8	97440	B/o Jothi	2/365	M	2.5	Ano cutaneous fistula			Anoplasty	low					Good	+
9	96355	Alagupandy	1	M	7	Recto prostratic fistula	Primary VUR Lt	TC	Sacro abdominal PT	low	Retraction, Redo, Skin erosion	Anal stenosis, Redo Ano plasty			Poor	+
10	99133	Thiva	8/12	M	8	Ano cutaneous fistula			Anoplasty	low					Good	+
11	98727	Selva Jothi	8/12	F	4.5	Ano Vestibular fistula			Post. Transposition	low		wound infection			Good	+
12	101583	Jeyapriya	2	F	6	Ano Vestibular fistula			Post. Transposition	low					Good	+
13	102105	Srikarani	6/12	F	6	Vulvar anus			Post. Transposition	low					Good	+
14	101176	Rahima	1 1/2	F	7	Anterior perineal anus	Hemi vertebra		Post. Transposition	low					Good	+
15	104256	Gunaseelan	3/365	F	3	Complete covered anus			Anoplasty	low		Stricture Redo			Good	+
16	106366	B/o Veeramm	2/365	M	1.7	ARM - High	OE & TEF, Hemi vertebra	TC	Thoractomy and Repair	low				Death		+

17	107508	Kannan	1	M	7	Ano cutaneous fistula			Anoplasty	low					Good	+
18	200782	B/o Sangeeth	2/365	F	2	ARM - Intermediate		TC		low				Death		+
19	24011	B/o Esakkiam	1/365	M	1.76	Ano cutaneous fistula			Anoplasty	low					Good	+
20	23198	Yasmitha	6/12	F	6.5	Cloacal anamoly	VSD, Absent S3, S4	TC	PSARP	low	Skin erosion				Poor	+
21	26749	Asarutheen	8/12	M	6	Recto vesicle fistula	VSD polydactily	TC	LAAPT	low	Prolapse, Skin erosion	Mucosal prolapse	Stitch granuloma		Fair	+
22	26439	Sangeetha	3.6/12	F	12	Ano Vestibular fistula			ASARP	low		Rectal stricture, Redo anal			Fair	+
23	29989	Divya Bharath	1	F	8	Anterior perineal anus	Hemi vertebra		ASARP	low					Fair	+
24	30407	Pavithra	2	F	15	Ano Vestibular fistula			Post. Transposition	low					Good	+
25	39289	Vasantha	8/12	F	5	Ano cutaneous fistula			Anoplasty	low		Revision Anoplasty			Good	+
26	41665	B/o Jayakodi	4/365	M	1.7	Recto vesicle fistula	TEF, PDA, Radial dysplasia	TC	TEF Repair & TC	low				Death		+
27	43802	Pandi Selvi	10	F	2.2	Ano Vestibular fistula	Primary VUR Rt		Post. Transposition	low					Good	+
28	43939	Harish	4/12	M	5.2	Ano cutaneous fistula	Patent Urachus		Anoplasty	low					Good	+
29	44363	B/o Arumugan	1/365	F	2	Complete covered anus			Anoplasty	low					Good	+
30	50410	B/o Isabella	3/365	M	2.5	Complete covered anus	Micro penis		Anoplasty	low					Good	+
31	41496	B/o Jeya	2/365	M	2.5	Complete covered anus			Anoplasty	low					Good	+
32	58304	Beaula	7/12	F	6	Ano Vestibular fistula	Mild Left HN		Post. Transposition	low		wound infection			Fair	+
33	60927	Mathana	3	F	11	Recto vesical fistula			Sacro abdominal PT	low					Fair	+
34	65902	Swetha	8/12	F	8	Recto vaginal fistula (low)	VSD	PC	LAAPT	low	Prolapse	Mucosal prolapse			Good	+
35	67537	Vidhya sree	2 1/2	F	7	Cloacal anamoly		TC	PSARP	low	Prolapse				Fair	+
36	71471	Yokeshwaran	8/12	M	7	Recto bulbar fistula	Mid penil HPS	PC	PSARP	low	Bleeding		wound infection		Good	+
37	73693	Pandi meena	6/12	F	7	Recto vaginal fistula(high)	TOF, Malrotation	TC	Lap. Sacro abdominal PT	low	Bleeding, Obstruction				Good	+

38	74282	Lingesh	11/12	F	6	Ano cutaneous fistula			Anoplasty	low					Good	+
39	77544	Anitha	8	F	15	Ano Vestibular fistula			Post. Transposition	low		Redo transposition			Poor	+
40	77232	Kabilan	4/12	M	5.5	Without fistula		PC	PSARP	low					Good	+
41	77356	Madan kumar	4	M	9	Recto bulbar fistula	Pelvic kidney	PC	PSARP	low	wound infection		Stitch granuloma		Good	+
42	79154	Vinoth kumar	2	M	10	Recto prostatic fistula	Hemi vertebra	PC	Sacro abdominal PT	low		wound dehesion			Good	+
43	82477	Velmurugan	1	M	7	Pouch colon	Absent 12th rib	END	Sacro abdominal PT	low	wound infection				Fair	+
44	83797	Kishore	10/12	M	7	Ano cutaneous fistula			Anoplasty	low					Good	+
45	86312	Kathirvel	1 1/2	M	10	Recto prostatic fistula	Polydactily	PC	PSARP	low	Skin erosion	Mucosal prolapse			Good	+
46	85675	Vijayalakshmi	1	F	5	Ano Vestibular fistula			Post. Transposition	low					Good	+
47	88613	Arthi	8/12	M	10	Ano cutaneous fistula			Anoplasty	low					Good	+
48	89270	Vasanthkuma	1 1/2	M	8	Recto vesicle fistula	Malrotation, Hemi vertebra	TC	Sacro abdominal PT	low	Skin erosion		wound infection		Fair	+
49	93211	B/o Ramya	1/365	M	2.5	Ano cutaneous fistula			Anoplasty	low					Good	+
50	90762	Pandi	10/12	M	6.2	Recto bulbar fistula		PC	LAAPT	low					Good	+
51	93862	Nilarishan	10/12	M	7	Ano cutaneous fistula	Hemi vertebra		Anoplasty	low					Good	+
52	97779	Priyadharshin	1	F	5	Ano Vestibular fistula			Post. Transposition	low					Fair	+
53	101249	B/o Amaravat	2/365	M	2	ARM - High	TEF Sacral agenesis, penoscrotal HPS	TC	TEF Repair & TC	low				Death		+
54	102570	Angamuthu	1	M	7	Recto bulbar fistula	12th rib Absent	PC	LAAPT	low	Skin erosion		obstructio n		Poor	+
55	1150	Thiruvan	6/12	M	9	Recto bulbar fistula	Lt crossed fused kidney	PC	PSARP	low		Mucosal prolapse			Fair	+
56	360	B/o Kavitha	1/365	F	2.7	Complete covered anus			Anoplasty	low					Good	+
57	9565	Akura	7/12	F	6.7	Anterior perineal anus			ASARP	low					Good	+
58	13705	Yogashri	10/12	F	6	Recto vaginal fistula (low)		PC	PSARP	low	Prolapse, redo	Retraction Redo Anorectoplasty	Stitch granuloma		Fair	+

59	16537	Aswin	10/12	M	6.4	Recto vesicle fistula	ASD, syndactaly	PC	Sacro abdominal PT	low	Bleeding		Leak		Good	+
60	18781	Kingsly	3	M	12	Ano cutaneous fistula			Anoplasty	low					Good	+
61	20120	Arunadevi	9/12	F	6	Vulvar anus			Post. Transposition	low					Good	+
62	241396	Mohammed	1	M	9	Recto prostratic fistula	PDA	PC	Sacro abdominal PT	low					Good	+
63	19986	B/o Vasuki	1/365	M	3	Complete covered anus			Anoplasty	low					Good	+
64	23760	B/o Muthamm	1/365	M	17	Ano cutaneous fistula	Bifid scrota		Anoplasty	low					Good	+
65	23978	Swetha	1	F	8	Recto vestibular fistula	Bladder diverticula	PC	PSARP	low	prolapse		infection, inc.hernia		Good	+
66	25101	Suguna	7	F	18	Ano cutaneous fistula			Anoplasty	low					Good	+
67	23920	Sandru	6/12	M	6	Recto bulbar fistula	Lt UDT	PC	PSARP	low		Fatal fistula			Poor	+
68	24693	Kalaiyarasi	6/12	F	7	Rectal Atresia		TC	PSARP	low	skin erosion		Leak		Good	+
69	25170	Sahana selvi	1 3 /12	F	6.5	Recto vaginal fistula (high)	Polydactily, hemi vertebra	TC	Sacro abdominal PT	low	Obst. Redo		Stitch granuloma		Good	+
70	27277	Pavithra	1 3 /12	F	9	Vulvar anus	Primary VUR		ASARP	low					Good	+
71	28827	Vinoth kumar	5	M	14	Ano cutaneous fistula			Anoplasty	low					Good	+
72	28434	Dhanusha	5	F	18	Anterior perineal anus			ASARP	low					Good	+
73	29248	Mohan	8/12	M		Recto bulbar fistula		PC	LAAPT	low	Skin erosion	Anal stenosis, Redo Anoplasty			Fair	+
74	32512	sivaprakash	5/12	M	5.3	Without fistula (I)	Down's syndrome	PC	PSARP	low					Good	+
75	39974	B/o Ribalya	1 /365	M	2.5	Ano cutaneous fistula			Anoplasty	low					Good	+
76	40533	Arunadevi	9/12	F	5	Recto vestibular fistula	Horse Shoe kidney	PC	PSARP	low		Retraction Redo Ano rectoplasty		Death		+
77	39855	Rupilan	2 6/ 12	M	10	Recto prostratic fistula		PC	Sacro abdominal PT	low					Poor	+
78	42746	B/o Tamilselv	1/365	M	2.5	Ano cutaneous fistula	Polydactily		Anoplasty	low		Stricture			Good	+

79	43068	Priyanka	1	F	5.2	Anterior perineal anus			ASARP	low					Good	+
80	45198	B/o Chellamm	3/365	M	2.9	Complete covered anus			Anoplasty	low					Good	+
81	46695	Sabari krishna	9/12	M	8	Recto vesicle fistula	Neurogenic bladder	PC	LAAPT	low	Retraction		wound infection		Good	+
82	97863	Dhayasri	6/12	F	5.5	Recto vaginal fistula (high)	Hemi vertebra	PC	Sacro abdominal PT	low					Fair	+
83	49939	B/o Vidhya	1/365	M	2.5	Complete covered anus			Anoplasty	low					Good	+
84	59118	Alagammal	1	F	5	Ano Vestibular fistula			Post. Transposition	low					Good	+
85	53668	B/o Nithya	1/365	M	1.7	ARM - High		TC		low				Death		+
86	63845	Bhuvaneshwa	10/12	F	5	Ano cutaneous fistula	ASD with PDA		Anoplasty	low					Good	+
87	67660	B/o Eswari	6/365	F	3.5	Ano cutaneous fistula			Anoplasty	low					Good	+
88	59895	B/o Vijaya	2/365	M	1.5	ARM - High	B/L Renal dysplasia, common AV channel	TC		low				Death		+
89	102543	B/o Rani	2/365	M	3	ARM - Intermediate	Meconium ileus	Ileos tomy		low				Death		+
90	86630	Siva	6/12	M	6	Rectal Atresia		TC	PSARP	low	Prolapse bleeding		Adhesive obstructio n		Good	+